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Truncus Arteriosus With Single Pulmonary Artery: Influence of Pulmonary Vascular Obstructive Disease on Early and Late Operative Results

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Nineteen patients with truncus arteriosus and single pulmonary artery had corrective operations at the Mayo Clinic from 1969 to 1983. At operation, their ages ranged from 4 months to 20 years (mean 8.1 years). The preoperative pulmonary resistance divided by 2 was used to predict the degree of pulmonary vascular obstructive disease at operation. The influence of elevated pulmonary resistance and the intraoperative postrepair ratio of pulmonary artery to left ventricular pressure on operative and late mortality were examined.

The outcome of patients with single pulmonary artery was compared with the outcome of 148 patients with truncus arteriosus and two pulmonary arteries operated on during the same period. Patients with a single pulmonary artery had an operative mortality similar to that of patients with two pulmonary arteries (32 versus 28%, $p > 0.05$). Late mortality was, however, significantly greater ($p < 0.001$) for patients with a single pulmonary artery. Elevated ratios of intraoperative postrepair pulmonary artery to left ventricular pressure were associated with significantly higher ($p < 0.02$) operative and late mortality, but elevated preoperative pulmonary resistance was not ($p > 0.10$).

Truncus arteriosus with single pulmonary artery is associated with poor postoperative survival, and although elevated pulmonary resistances preoperatively did not predict outcome, elevated intraoperative postrepair pulmonary artery to left ventricular pressure ratios were associated with increased operative and late survival, suggesting a deleterious role of pulmonary hypertension.

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The natural history of uncorrected truncus arteriosus is characterized by the inevitable development of pulmonary vascular obstructive disease in those patients who survive infancy (1). Obstruction follows prolonged hyperperfusion of the pulmonary arteries at levels of systemic pressure. The pulmonary vascular obstructive disease may be exacerbated further in patients with either congenital absence or acquired occlusion of a pulmonary artery (2).

Greatly elevated pulmonary vascular resistance in patients with truncus arteriosus and two pulmonary arteries has been reported to increase the surgical mortality (3,4). Postmortem histologic examination of the pulmonary vascular bed from such patients shows good correlation between an elevated pulmonary resistance and the degree of pulmonary vascular obstructive disease.

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Previous investigators (4) noted that patients with truncus arteriosus and two pulmonary arteries had high operative mortality when the total pulmonary resistance before operation was greater than 8 $U \cdot m^2$. Mair et al. (3,5) reported that, in patients with truncus arteriosus and a single pulmonary artery, the most reliable predictor of the degree of pulmonary vascular obstructive disease was half rather than total pulmonary resistance.

This study examines the long-term results of intracardiac repair of truncus arteriosus with single pulmonary artery and evaluates the influence of elevated preoperative pulmonary resistance on survival and the occurrence of pulmonary vascular obstructive disease. Operative and late survival data also were compared with data from patients with truncus arteriosus and two pulmonary arteries who were operated on at the Mayo Clinic during the same period.

Methods

Patient description. At the Mayo Clinic from 1969 to 1983, 19 consecutive patients with truncus arteriosus and

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single pulmonary artery had intracardiac repair as described previously (6). Their ages ranged from 4 months to 20 years (mean 8.1 years). In 16 of the 19 patients, one pulmonary artery was congenitally absent; 3 of the 16 had previous banding of the single pulmonary artery at 2 months, 10 months and 11 years of age, respectively. In the three other patients, one pulmonary artery was completely occluded from previous pulmonary artery banding performed at 2½, 6 and 9 months of age, respectively. Patients with segmental proximal atresia or occlusion of one pulmonary artery in whom continuity with a normal-sized distal pulmonary artery was surgically reestablished were excluded.

Preoperative and intraoperative evaluation. All patients were evaluated preoperatively by cardiac catheterization to determine hemodynamic criteria for operability based on an assessment of the total pulmonary resistance, that is, the mean pulmonary artery pressure divided by the pulmonary flow index. Pulmonary flow indexes were measured by the Fick method. Operation was offered only to patients with truncus arteriosus and single pulmonary artery in whom half of the pulmonary resistance was 8 U·m^2 or less while the patient was breathing room air or 100% oxygen (5). Intraoperative postrepair measurements of peak systolic pulmonary artery (16 patients) or right ventricular (3 patients) to left ventricular pressure ratios were compared as indicators of elevated pulmonary artery pressure after operation. These data were compared with data from 148 patients with truncus arteriosus and two pulmonary arteries who were operated on during the same period.

Follow-up. All surviving patients completed a written questionnaire that evaluated their clinical status. Information also was obtained by review of the details of their most recent clinical evaluations, either at our institution or by their personal physician.

Statistical methods. Relation of continuous variables and hospital mortality were assessed by the two-tailed Wilcoxon rank-sum test. This nonparametric two-sample test was used instead of the *t* test because of the highly skewed distribution of values for preoperative pulmonary resistance. Associations between late mortality and hemodynamic variables were assessed using the log-rank transformation (7) and applying the Cox proportional hazards model (8). Statistical comparisons between the unilateral and bilateral pulmonary artery groups were based on the chi-square test for hospital mortality and the log-rank test for long-term mortality.

Results

Pulmonary resistance and pressure. In 18 of the 19 patients operated on (Table 1), the preoperative total pulmonary resistance ranged from 3.4 to 18.4 U·m^2 (mean 10.7). When these values are expressed as pulmonary resistance divided by 2, the resistance values ranged from 1.7 to 9.2 U·m^2 (mean 5.3). In 1 of the 19 patients (Case 1), pulmonary resistance in room air was 33.6 U·m^2 but decreased to 16.4 U/m^2 (8.2 pulmonary resistance ÷ 2) during the breathing of 100% oxygen. The intraoperative postrepair pulmonary artery to left ventricular pressure ratios of 16 patients whose pulmonary artery pressure was measured ranged from 0.31 to 1.2 (mean 0.64). In the other three patients, the right ventricular to left ventricular pressure ratios were 0.57, 0.60 and 0.60, respectively.

Perioperative deaths. Six (32%) of the 19 patients died at or within 1 month of operation. Autopsy was performed in five of the six. In four of the six (Cases 2, 4, 7 and 9), pulmonary resistance was elevated, having values of 12.3 to 18.4 U·m^2. Histologic changes of irreversible pulmonary vascular obstructive disease (Heath-Edwards grade ≥3) (9) were present at autopsy in the lungs supplied by the single pulmonary artery in three of the four patients. In the fourth patient (Case 9), who had a preoperative pulmonary resistance of 12.3 U·m^2, only grade 1 Heath-Edwards lesions were found at autopsy; in this case, hypoplasia of the distal pulmonary artery probably led to overestimation of the preoperative pulmonary resistance and to elevated pulmonary artery to left venticular pressure ratios (1.2) after repair. Autopsy in the fifth patient (Case 16), who had a low preoperative resistance (5.4 U·m^2), revealed only grade 1 Heath-Edwards changes in the pulmonary vascular bed supplied by the single pulmonary artery.

Preoperative elevation of pulmonary resistance was unassociated with a higher operative mortality rate. The mean pulmonary resistance was 11.5 U·m^2 for the 5 patients who died at operation and 10.3 U·m^2 for the 13 survivors. However, the intraoperative postrepair pulmonary artery to left ventricular pressure ratio was significantly higher in patients who died at operation than in survivors. The mean intraoperative postrepair pulmonary artery to left ventricular pressure ratio for survivors was 0.55 compared with 0.73 for patients who died at operation ($p < 0.02$). The overall operative mortality rate of 32% for patients with truncus arteriosus and a single pulmonary artery was not significantly different from the rate of 28% for patients with truncus arteriosus and two pulmonary arteries.

Late deaths. Seven late deaths (Cases 1, 5, 6, 8, 10, 12 and 14) occurred from 18 to 100 months (mean 63.6) after operation. The preoperative pulmonary resistance ranged from 7.4 to 33.6 U·m^2 (mean 15.0). In three of these seven patients, severe pulmonary vascular obstructive disease was demonstrated either by autopsy (Cases 6 and 10) or by documentation of severe pulmonary hypertension at late recatheterization (Cases 5 and 10). In Cases 1 and 8, autopsy was not done. The remaining two patients had no evidence of severe pulmonary hypertension or plexogenic pulmonary vascular obstructive disease by late recatheterization (Case 12) or autopsy (Case 14). However, one of the patients (Case 14) had evidence of thromboembolic pulmonary vascular disease. Although the preoperative pulmonary resist-

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Table 1. Outcomes of 19 Patients With Truncus Arteriosus and Single Pulmonary Artery

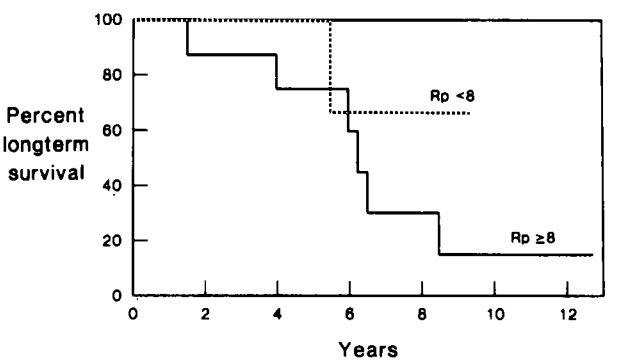
Case	Age at Operation (yr)	Previous PA Banding (age)	Preop Rp (U·m²)	Intraoperative Postrepair Systolic Pressure: Distal PA Pressure (mm Hg)	Intraoperative Postrepair Systolic Pressure: Systemic LV or Aortic Pressure (mm Hg)	PA/LV	Follow-up and Present Status	Outcome*
1	11	None	33.6 (16.4)†	60‡	90	0.66§	Late death, 18 mo postop	No autopsy
2	20	11 yr	18.4	90	100	0.90	Operative death	Gr 3 lesions at autopsy
3	7	None	17.1	62	115	0.54	Alive, 147 mo postop	Catheterization 9 yr postop; PAP 70 mm Hg
4	1 5/12	None	17.0	75	85	0.88	Operative death	Gr 4 lesions at autopsy
5	8	2 mo	16.0	45	105	0.43	Late death, 100 mo postop	Catheterization 3 yr postop; PAP 120 mm Hg
6	15	None	15.0	75	105	0.71	Late death, 74 mo postop	Gr 5 lesions at autopsy
7	3	None	13.3	65	85	0.76	Operative death	Gr 4 lesions at autopsy
8	6	None	13.0	65	100	0.65	Late death, 70 mo postop	No autopsy
9	13	None	12.3‖	85	70	1.20	Operative death	Gr 1 lesions at autopsy
10	8	6 mo¶	11.6	70	110	0.64	Late death, 45 mo postop	Catheterization 3 yr postop; PAP 148 mm Hg, LV 90 mm Hg; gr 4 lesions at autopsy
11	5	None	9.3	60	110	0.55	Alive, 79 mo postop	—
12	10	10 mo	8.5	40	100	0.40	Late death, 77 mo postop	Catheterization 5 yr postop; PAP 4 2/12 mm Hg
13	6	None	7.8	30	80	0.37	Alive, 13 mo postop	Gr 1 and 2 lesions by biopsy at operation
14	11	9 mo¶	7.4	85	100	0.85	Late death, 61 mo postop	Thromboembolic pulmonary obstructive disease at autopsy
15	2	None	7.1	50	90	0.55	Alive, 74 mo postop	—
16	5	2½ mo¶	5.4	30	60	0.50	Operative death	Gr 1 lesions at autopsy
17	12	None	5.3	25	80	0.31	Alive, 133 mo postop	Catheterization 112 mo postop; Rp 3.3 U·m²
18	12	None	3.7	55‡	95	0.57§	Alive, 60 mo postop	—
19	5/12	None	3.4	45‡	75	0.60	Operative death	No autopsy

*Heath-Edwards grade: evidence of pulmonary vascular obstructive disease. †On 100% oxygen. ‡Right ventricular pressure rather than pulmonary artery pressure. §Right ventricular/left ventricular rather than pulmonary artery/left ventricular. ‖Preoperative total pulmonary resistance probably overestimated because of distal pulmonary artery stenosis. ¶Acquired pulmonary artery occlusion. Gr = Heath-Edwards grade; LV = left ventricle; PA = pulmonary artery; PAP = pulmonary artery pressure; postop = postoperative; Rp = total pulmonary resistance; RV = right ventricle.

ance tended to be higher in patients who died late (six of seven having a pulmonary resistance >8 U·m²) than in survivors (two of six of whom had pulmonary resistance >8 U·m²), this was not statistically significant because of the unequal times of follow-up in patients from each group (Fig. 1). Intraoperative postrepair pulmonary artery to left ventricular pressure ratios were, however, significantly higher in patients who died late than in survivors ($p < 0.02$) (Fig. 2). Late death was due to congestive heart failure with pulmonary hypertension in three patients and to sudden death in four patients, one of whom was receiving antiarrhythmic therapy. Surgical repair was intact in all autopsy cases.

Late survivors. Six (32%) of the 19 patients (Cases 3, 11, 13, 15, 16 and 18) were alive from 13 to 147 months (mean 84.3) postoperatively. The preoperative pulmonary

Figure 1. Long-term survival after operation for truncus arteriosus with single pulmonary artery: patients with preoperative pulmonary resistance (Rp) 8 U·m² or less compared with patients with pulmonary resistance greater than 8 U·m².



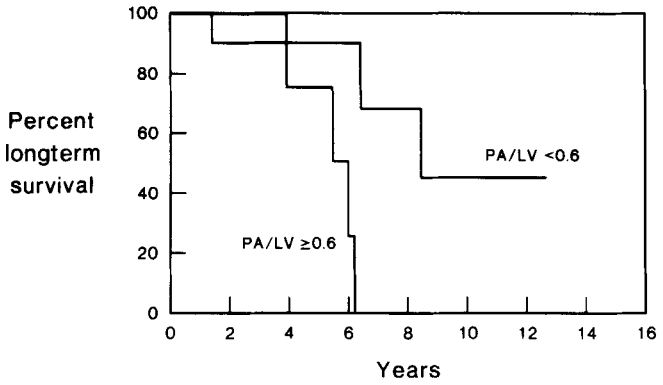


Figure 2. Long-term survival after operation for truncus arteriosus with single pulmonary artery: patients with intraoperative postrepair pulmonary artery to left ventricular (PA/LV) pressure ratio 0.6 or less compared with patients with pulmonary artery to left ventricular pressure ratio greater than 0.6.

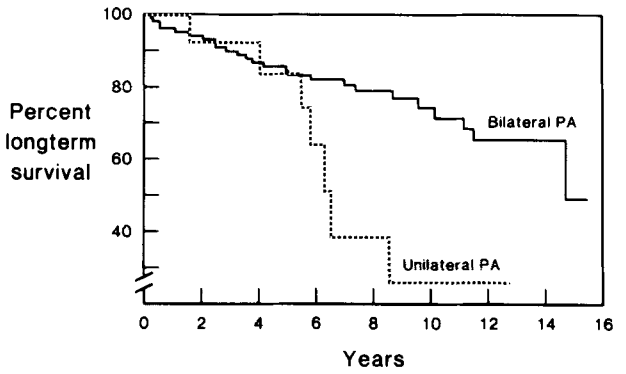


Figure 3. Long-term survival after operation for truncus arteriosus: patients with unilateral pulmonary artery (PA) compared with patients with bilateral pulmonary arteries.

resistance ranged from 3.4 to 17.1 U·m^2 (mean 7.8). Only one late survivor (Case 3) had evidence of pulmonary hypertension at catheterization, having a mean pulmonary artery pressure of 70 mm Hg, which has been unchanged in the 9 years since operation. One other patient (Case 17) has been recatheterized 112 months postoperatively and has a normal pulmonary resistance of 3.3 U·m^2.

Pulmonary vascular obstruction. When preoperative total pulmonary resistance was greater than 8 U·m^2 (pulmonary resistance ÷ 2 > 4 U·m^2), histologic evidence of advanced pulmonary vascular obstructive disease (Heath-Edwards grade 3 or 4) was present at autopsy in three patients who died at operation. Patients with a preoperative pulmonary resistance less than 8 U·m^2 had minimal histologic evidence of pulmonary vascular obstructive disease at operation (two patients by autopsy [Case 16] or lung biopsy [Case 13]) or had minimal pulmonary hypertension at late recatheterization (one patient [Case 12]). Long-term survival was slightly better in patients with pulmonary resistance less than 8 U·m^2 and was significantly better in patients with lower intraoperative postrepair pulmonary artery to left ventricular pressure ratios.

Late survival: one versus two pulmonary arteries. Late outcome of the 13 operative survivors with a single pulmonary artery was compared with late outcome of 106 operative survivors with truncus arteriosus and two pulmonary arteries who were operated on during the same period. The long-term outcome was significantly worse for the patients with a single pulmonary artery ($p < 0.001$) (Fig. 3).

Pulmonary artery banding. Prior pulmonary artery banding resulted in occlusion of a pulmonary artery in three of the patients in this series (Cases 10, 14 and 16). Of the six patients who had banding to one or both pulmonary arteries, two (Cases 5 and 10) still developed pulmonary hypertension.

Discussion

This study demonstrates that patients with truncus arteriosus and only a single pulmonary artery are at high risk for operative and late mortality and have a significantly reduced late survival compared with patients with truncus arteriosus and two pulmonary arteries.

Previous studies. In a previous study, Marcelletti et al. (4) demonstrated that patients with truncus arteriosus and two pulmonary arteries who had total pulmonary resistance greater than 8 U·m^2 had a worse operative outcome than did patients with pulmonary resistance less than 8 U·m^2. Although an elevated pulmonary resistance did not significantly predict a higher operative mortality in patients with a single pulmonary artery, an elevated intraoperative postrepair pulmonary artery to left ventricular pressure ratio was associated with both higher operative and late mortality. This strongly implicates pulmonary vascular obstructive disease as a significantly detrimental factor in operative and late survival.

Pulmonary artery banding often was ineffective in preventing pulmonary vascular obstructive disease and postoperative mortality in these and other patients (10). Pulmonary banding itself has been associated with an operative mortality rate of as high as 60% (11). Pulmonary banding also creates the potential hazard of migration of the band, causing occlusion of a pulmonary artery, as in three cases in this study. Intracardiac repair in infancy without prior banding avoids these hazards.

Significance of pulmonary resistance. In this study, long-term survival was significantly worse in patients with a single pulmonary artery than in cohorts with two pulmonary arteries. Patients with pulmonary obstructive dis-

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ease before operation may be at a greater risk of continued progression of the process than are patients with truncus arteriosus and two pulmonary arteries. This may be due to the entire cardiac output being directed through half the normal number of pulmonary arterioles, causing each arteriole to be subjected to twice the normal flow, which may predispose to progression of pulmonary vascular obstructive disease.

Although intraoperative postrepair pulmonary artery to left ventricular peak systolic pressure ratios correlated with operative and late mortality, preoperative pulmonary resistances did not have statistically significant predictive value on outcome. Although elevated pulmonary resistance did not consistently predict intraoperative postrepair pulmonary artery to left ventricular pressure ratios, pulmonary resistance of greater than 8 $U \cdot m^2$ (pulmonary resistance $\div$ 2 > 4 $U \cdot m^2$) was associated with histologic evidence of advanced pulmonary vascular obstructive disease. An error in estimation of pulmonary flow could occur if significant collateral blood supply was present to the lung of which the pulmonary artery was absent. This would cause calculated pulmonary flow to be overestimated and, therefore, pulmonary resistance to be underestimated. It may be prudent, therefore, not to rely on preoperative pulmonary resistance as a single criterion for operability in patients with truncus arteriosus and single pulmonary artery, when pulmonary vascular obstructive disease is considered likely. In older patients, a lung biopsy may be necessary to determine the presence or absence of severe pulmonary arteriopathy. Even when there is biopsy evidence of relatively mild pulmonary vascular obstructive disease, the potential for its postoperative progression and a poor late result should be explained to patients.

Early repair advocated. Advanced pulmonary vascular obstructive disease has been documented (12,13) in patients who are younger than 1 year of age with truncus arteriosus and two pulmonary arteries and was present in a 16 month old child (Case 4) in this series. This early development of irreversible pulmonary vascular disease in patients with truncus arteriosus suggests that complete repair should be attempted early in the first year of life in an attempt to prevent the development of pulmonary vascular obstructive disease. If only a single pulmonary artery is present, pulmonary obstructive disease may develop even more rapidly, and operation should be performed within the first few months of life. Recent reports (12–17) of successful correction of truncus arteriosus in infancy, with an acceptably low mortality rate, have made this approach to management of these patients feasible.

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